EJVES Extra **10**, 24–26 (2005)
doi:10.1016/j.ejvsextra.2005.04.012, available online at http://www.sciencedirect.com on SCIENCE DIRECT®

SHORT REPORT

Metachronous Haemangiopericytomas: Rare Vascular Entity

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Haemangiopericytomas is a rare vascular tumour. The trunk and lower extremities are involved in most cases. Fifteen to twenty percent of haemangiopericytomas arise in the head and neck.
We describe a case of a benign metachronous haemangiopericytoma in a 33-year-old man. The lesions were dealt by prompt surgical excision. This case highlights the need of close long-term follow-up of such patients with haemangiopericytoma even when benign.

Keywords: Haemangiopericytoma; Metachronous; Magnetic resonance imaging; Immunohistochemistry; Surgical excision.

Introduction

Haemangiopericytoma is an uncommon mesenchymal tumour which comprises of 1% of all blood vessel-related neoplasms and approximately 3% of all soft tissue sarcomas.[1] Being uncommon, haemangiopericytomas may be difficult to diagnose. About 15–25% of haemangiopericytomas arise in the head and neck.[1] In 1942, Stout and Murray coined the term 'haemangiopericytoma' for a unique vascular tumour which was characterised by branching capillaries surrounded by a reticulin network.[2] It presents as a painless mass, affecting males and females equally. The aetiology is unknown, although the presence of haemangiopericytoma has been linked to trauma and hormonal imbalance. The true nature of this neoplasm is uncertain and its clinical behaviour difficult to predict. Accurate diagnosis of haemangiopericytoma is made on the basis of conventional histological along with the use of immunohistochemical staining techniques with factor VIII-R-Ag, factor XIII and histocompatibility antigen HLA-DR.[3] The treatment of choice for haemangiopericytoma in any location is wide surgical excision.[1]

Case Presentation

A 33-year-old Caucasian male presented with a 1-year history of a painless nodular swelling on the right side of the neck which was associated with tingling on the right side of the face. Clinical examination revealed a tender $3\times3.2\ cm^2$ mass in the posterior triangle of the neck covered by freely movable normal skin. There had been no increase in the size of the swelling. There was no evidence of any enlarged lymph nodes. Technetium bone scan showed increased uptake in the lateral process of C2. The lesion was excised and macroscopic appearance consisted of a pseudoencapsulated mass measuring $3\times3.5\ cm^2$ in diameter. A portion of the lesion was solid but much of it was cystic with haemorrhagic necrosis. Histology demonstrated a cellular tumour with an intricate vascular network surrounded by polygonal cells in a typical staghorn a pattern and the excision seemed complete. Immunohistochemical staining with factor XIIIa demonstrated staining of the vascular spaces and the pericytic membranes (Fig. 1). The histological and immunohistological profiles were consistent with a benign haemangiopericytoma, with infrequent

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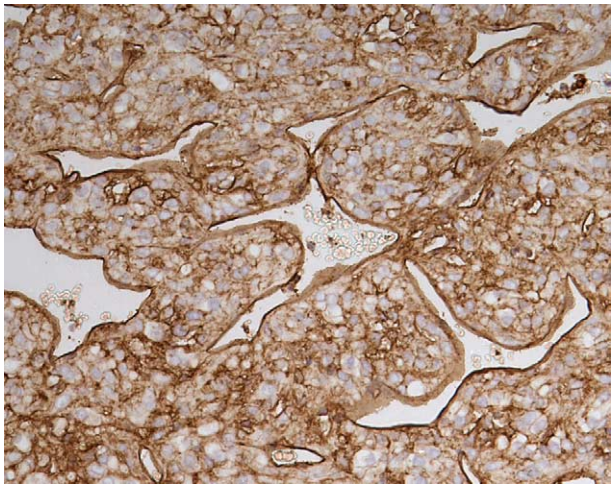

Fig. 1. Immunohistochemical staining with factor XIII shows intense staining of the vascular spaces and the pericytic membranes.

mitoses. Four years from the initial presentation, he presented with a firm mass in the left side of the neck. Magnetic resonance (MR) scan demonstrated a well-circumscribed, lobulated lesion on two-dimensional images. Furthermore, it revealed a non-uniform high signal intensity with increased vascularity on T2-weighted images without infiltration of the surrounding tissue (Fig. 2). The lesion was excised and the histological evaluation proved to be haemangiopericytoma. In the following year, he was seen in the clinic with a firm $3\times4\,cm^2$ lump in the right thigh. There was no evidence of distal neurovascular deficit. The lesion was excised and the histology proved to be

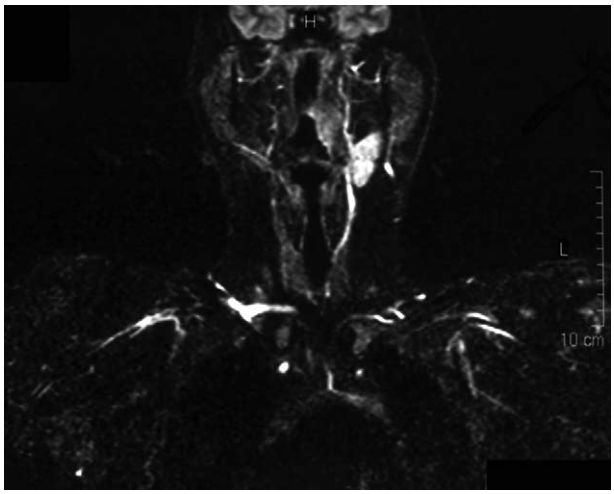

Fig. 2. Magnetic resonance (MR) scan demonstrates a well-circumscribed, lobulated, lesion with non-uniform high signal intensity with increased vascularity on T2-weighted images without infiltration of the surrounding tissue.

haemangiopericytoma again without any overt features of malignancy. The patient has remained well with no further lumps in the subsequent 6 months follow-up.

Haemangiopericytoma is an uncommon vascular tumour that may pose difficulty in diagnosis. Like the glomus tumour, haemangiopericytoma arises from capillary pericytes; for this reasons it may arise in any part of the body where capillaries are present. Pericytes are slender, elongated cells located in the walls of capillaries and venules.[4] They are external to the endothelial cells and are separated by a basement membrane. Whereas, haemangiopericytomas commonly arises in the retroperitoneum and lower extremities, about 15–25% of them arise in the head and neck region.[1] Haemangiopericytoma has no race predilection and it has been found in patients of all ages. It presents as a slow-growing, painless mass that is often nodular and well circumscribed. The overlying skin or mucosa is usually normal in appearance. The diagnosis of this tumour can be problematic. The differential diagnosis includes lymphomas, neurofibromas, fibrosarcomas and solitary fibrous tumours. Imaging, biopsy and immunohistochemical staining are essential for establishing the diagnosis.

Haemangiopericytomas have higher potential of relapse, local invasiveness, and the possibility of producing distant metastases. Magnetic resonance angiography is a better diagnostic method than a computed tomographic scans for this vascular tumour, which usually demonstrates hypervascularity. Microscopically the tumour is composed of a large number of capillaries and small vessels, dense spindle-shaped tumour cells, and reticular fibres. Imunohistochemically, haemangiopericytomas are known to show a positive response to antibodies against vimentin and type IV collagen, factor XIII and a negative response to VIII-related antigen, S-100 protein, neuronspecific enolase carcinoembryonic antigen, desmins, laminin, and cytokeratins.[3]

Although distinction between benign and malignant haemangiopericytoma cannot be made in all cases, careful evaluation of certain morphologic criteria is helpful in predicting the clinical course. Prominent mitoses, necrosis, haemorrhage, and increased cellularity often associated with thrombosis are features usually observed in tumours that later recur or metastasise.[5]

Because of the rarity and unpredictable biologic behaviour of these neoplasms, there is controversy about the best way to manage them. It is generally accepted that the treatment of choice is radical surgical excision when the tumour is localized and technically resectable. Radiotherapy is recommended in cases of

incomplete tumour removal or of unresectable and recurrent lesions or for palliation. The combination of radical surgery, postoperative radiation therapy, and close follow-up represents the most ideal approach for locally aggressive, non-metastatic haemangiopericytoma. The wide *en bloc* surgical resection may be facilitated if the afferent vessels are preoperatively embolised to decrease tumour bleeding during surgery. Because of the high incidence of local recurrence, long-term follow-up is essential. In the present patient, the resected surgical margins were free of tumour and, therefore, no adjuvant radiotherapy was necessary in all the three sites.

This case report emphasises that haemangiopericytoma may be metachronous. In this case, there were no features of malignancy and excision margins were uninvolved. Despite this the patient had more lesions later on. Therefore, we feel that close long-term follow-up is essential not only for malignant haemangiopericytoma, but also for patients with benign haemangiopericytomas with uninvolved excision margins.

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Accepted 27 April 2005